CASE REPORT

FEMALE EPISPADIAS: SPOT DIAGNOSIS AND SIMPLE RECONSTRUCTION

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Isolated female epispadias without bladder exstrophy is a rare congenital anomaly. Patients present with total or partial urinary incontinence. The diagnosis can only be made on careful genitalia examination by separating the labia majora. That is why it is often missed by most physicians even after being extensively investigated. The physical findings include patulous urethra, flattened mons pubis, and bifid clitoris with lack of anterior labial commissure. In most of the cases, single stage reconstruction of urethra, labia minora and clitoris is enough to achieve urinary continence with cosmetically acceptable genitalia.

Keywords: Female epispadias, urinary incontinence, single stage repair

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INTRODUCTION

Female epispadias without bladder exstrophy is a very uncommon congenital anomaly occurring in one in 484,000 female births.[1] These girls usually present, at the age of toilet training, with urinary incontinence varying from continuous dribbling of urine to few episodes of day-time wetting.

The diagnosis is often missed unless carefully examined the genitalia. The characteristic appearance of genitalia is bifid clitoris, flattened mons pubis, poorly developed labia, patulous urethral meatus and sometimes symphyseal separation. The upper renal tract, vagina, and internal genitalia are usually normal in these patients.[2–4]

We are presenting this case firstly because it is a rare case and secondly to emphasise upon careful genitalia examination of an incontinent child to make early diagnosis of female epispadias and to prevent these girls from a prolonged misery of urinary incontinence.

CASE REPORT

A 6-year-old girl presented with dribbling of urine day and night since birth. She would void a scanty amount of urine on desire with a poor stream but would start dribbling after about half an hour. There was no other urological complaint. She was constipated most of the time. She had devastated social history and even at this age would prefer to remain isolated.

She had been extensively investigated in the past with no conclusion. Her renal function tests were within normal limits. Ultrasonography showed normal upper tracts with reasonable bladder capacity of 187 ml. Radiograph of the pelvis didn't show diastases. An improperly performed voiding cystourethrogram revealed small capacity bladder with bilateral grade-II vesico-ureteric reflux.

Examination of external genitalia revealed ill developed labia minora with widely separated two halves of the clitoris. The urethral meatus was wide and patulous. Vagina was normal (Figure-1).

Her surgery was planned. In lithotomy position an inverted Y shaped incision was given starting from mons pubis and inferiorly the limbs of Y extended on either sides of urethra on to 3 and 9'0 clock positions. Anterior wall of urethra mobilized in this plan all the way up to the bladder neck underneath the symphysis pubis.

A wedge of redundant urethra was excised and urethra was reconstructed over 10 Fr silicon catheter. Skin from the medial aspect of the hemi-clitoris and labia minora were denuded and clitoral reconstruction was done by approximating the corporal tissues.

Mons pubis reconstructed by undermining fibro fatty tissue and closing it in two layers after lateral mobilization to obliterate the dead space. This manoeuvre produced an excellent good cosmetic appearance (Figure 2–6).

She was discharged on next day on oral oxybutynin, paracetamol and antibiotics. Urethral catheter was removed after 7 days. She is dry now with significant urinary frequency in the initial days till reasonable bladder did not achieve.

DISCUSSION

Female epispadias is a rare congenital anomaly with an incidence of 1/484,000 female births.[1] The associated incidence of vesicoureteric reflux is reported to be 30–75%.[2] Urinary incontinence is invariably present at presentation but intensity varies depending upon functional bladder capacity. The

condition often gets attention at the age of toilet training when the child still wets. Urinary incontinence has serious social and psychological consequences and demands early management. The diagnosis is very simple by finding characteristic bifid clitoris, flat mons pubis, ill developed labia minora and wide patulous urethral meatus. Diagnosis is often missed when proper examination of genitals is not carried out by separating the labia majora. Upper urinary tract, vagina and internal genitalia are usually normal.[2–4] Reconstructive technique is simple in most of the cases. The single stage repair has good and acceptable appearance of external genitalia with 60–87% urinary continence rate.[5,6]

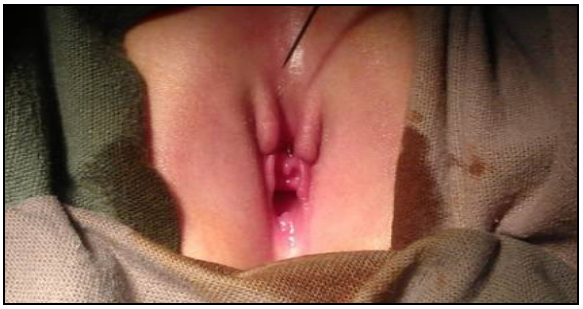
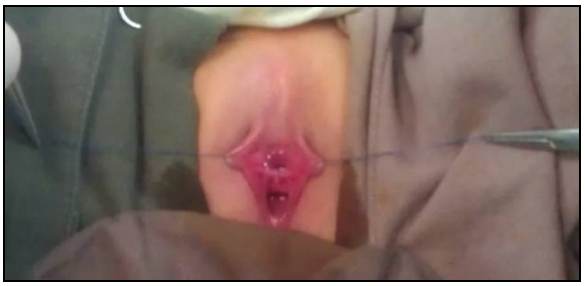


Figure-1



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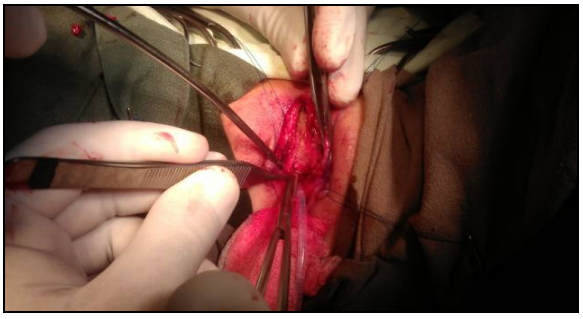


Figure-2

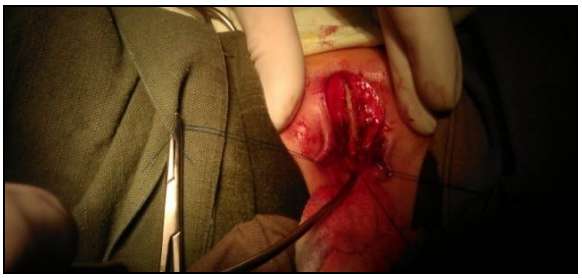
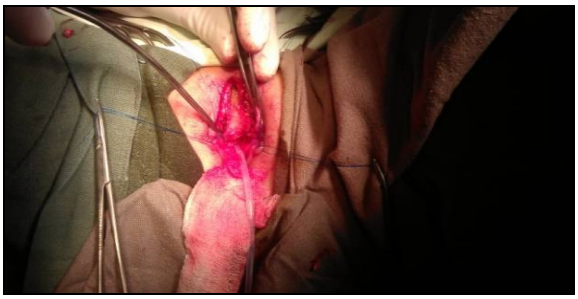


Figure-3



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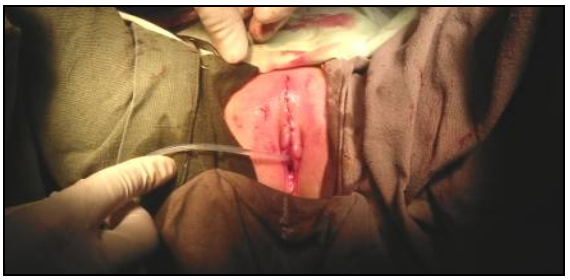

Figure-6

REFERENCES

1. Atilgan D, Uluocak N, Erdemir F, Parlaktas BS. Female epispadias: a case report and review of the literature. Kaohsiung J Med Sci 2009;25(11):613-6.
2. Gearhart JP, Mathews R. Exstrophy-Epispadias Complex. In: Wein AJ, editor. Campbell-Walsh Urology. 9th ed. Philadelphia: Saunders, 2007;p. 3547-50.
3. Mollard P, Basset T, Mure PY. Female epispadias. J Urol 1997;158:1543–6.
4. Pelzer AE, Akkad T, Schwentner C, Strasser H, Bartsch G, Rehder P. Treatment of adult female epispadias without exstrophy in the presence of rhabdosphincter function. Int J Urol 2006;13:321–2.
5. Gearhart JP, Peppas DS, Jeffs RD. Complete genitourinary reconstruction in female epispadias. J Urol 1993;149:1110–3.
6. Hendren WH. Congenital female epispadias with incontinence. J Urol 1981;125:558–64.

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